

## Hare Lip and Cleft Palate

By G. D. F. McFADDEN, M.B., M.CH.(BELF.), F.R.C.S.(ENG.)

"A BEAUTIFUL FACE is a silent commendation," wrote Francis Bacon, and he might with equal truth have written, "an ugly face is a silent condemnation." A baby born with a hare lip must produce almost a feeling of revulsion in the mother and relatives. It takes a great mother-love to bring up with affection and care this almost hideous progeny. The condition occurs about one in every thousand births, so that there are probably thirty such infants born every year in Ulster.

On account of their appearance, these children are hidden from the public view. They are not wheeled out to the fresh air; they get little sunshine. This, coupled with the difficulties and laboriousness of feeding, causes many of them to fail in health. Some die; others are so feeble when first seen that there is no question of operation till their general health is improved. One child in this series only weighed  $7\frac{1}{2}$  lb. when three months old. It took another three months to make this infant fit for operation.

The hare lip may be complete or incomplete and be unilateral or bilateral (see Plates A). The defective lip may be continuous, with a complete cleft of the palate, or the palate may be normal or only partially cleft, or the lip may show a dent. Probably the most difficult lips to rectify are those where the palate is completely united. It is easiest to get a good result in a lip in which the nostrils are not affected. Hare lip in a man may be worse than in a woman, for the central area of the cleft is devoid of hair and the cleft is exaggerated. The growth of hair is associated with muscle fibre<sup>1</sup> (fig. V. C.).



V. C. (After Veau)

This patient has had a right unilateral hare lip. Note that the medial side of the cleft is sterile, being devoid of hair.

There are two embryological areas involved: an anterior part, associated with hare lip, and a posterior part, associated with cleft palate.

The defect giving rise to hare lip is the result of a lateral defect, just as a cleft palate is the result of a median defect. Note in the complete facial cleft (Plate A1) the palate is not affected. The defect must take place about the sixth to seventh week in intrauterine life. In the embryo there is no red margin to a lip until the muscle fibres grow into it. Where muscle is defective, the red margin will be defective.

Although there is a wide cleft, all the elements going to form the lip, or the palate, are present—as yet undeveloped and possibly misplaced. I think a law can be formed, stating that in cases of congenital dehiscence in any part of the body the elements are all present; although they may be turned aside or undeveloped—they are never absent. The surgeon in hare lip and cleft palate aims at bringing these embryological elements into their natural place, so that as growth occurs the normal will be produced. A plastic surgeon might use flaps from other areas to fill the gap, and for the moment produce an improved appearance, but, as growth takes place, the abnormal will become exaggerated and the lip “be a reproach to the surgeon and an ever recurring reminder to the patient of a surgical barbarity.” As Veau states, we operate upon a face undergoing evolution—all the anatomical elements are indispensable to make sure of equilibrium in formation. If, unfortunately, one of these anatomical elements is misplaced, the equilibrium is broken and evolution adapts itself to this new state of dynamics, so that growth produces necessary new shapes which are essentially ugly, for the eye can only find beauty in the normal.

These embryological facts are the basis of modern treatment of hare lip and cleft palate. It was neglect of these that made the classical operations of Rose, Mirault, Jalaguier, Koenig, and so-called plastic operations such a failure (Plate D). Yet it seems strange that in one of the most recent books on plastic surgery I find only these operations recommended, and in one of the most popular of recent textbooks of surgery and one on dental surgery only those operations that have been found wanting and are outmoded are illustrated.

In a complete unilateral hare lip with cleft palate the whole bony skeleton of the face is altered. The nasal septum is drawn to one side and the maxillary bones underlying are deformed (Plate E), so that no operation on the soft parts can wholly correct the asymmetry. Attempts at breaking the bone or repositioning the bones have only been met with disaster. The only way is to allow time, acting through the corrected soft parts, to mould the bones into a more normal shape.

When the hare lip is being repaired in a complete cleft associated with cleft palate, it is essential to close the anterior part of the palate and to form a good floor for the nostrils. For, if the hare lip is repaired without the floor of the nostril and anterior part of the palate being reformed, it will be impossible to get at these areas at a later date, leaving a resultant chronic fistula (Plate D). By repairing the floor of the nostril and the anterior part of the palate the closure of the rest of the palate is made more simple at the second operation.



**Fig. E**

Drawn from a skeleton of a case with unilateral complete cleft palate. It shows a marked distortion of the nasal septum.

*(After Veau, with kind permission of Messrs. Masson et Cie)*

In double hare lip, where the premaxilla is completely separate (Plate A3) and carried forward, it is unwise to do both sides at the same operation. For the bilateral stripping of the mucous membrane so depletes the blood supply that gangrene and sloughing of the central premaxilla may result. In these cases one side is first done, so converting a double hare lip to a single hare lip, and about two months later the other side is repaired.

Lately Denis Browne has described an operation in double hare lip in two stages that promises to be an improvement on the older methods (Plate T).

For the modern operations on hare lip we owe much to Veau,<sup>1</sup> and I make no excuse for quoting freely from his book.

There are four main points in the operation :—

1. The surroundings must be freely mobilised.
2. The natural elements must be brought into their natural site. Here muscle must be sought and brought across to meet muscle.

3. The red margins must be accurately opposed.
4. The columella must be respected.

Every good lip is thick and pliable—full of muscle. This muscle grows from the lateral margin of the cleft and is the guardian of the red mucous membrane. The internal flap is sterile (see fig. V. C.), being devoid of hair and muscle. The mucous membrane on the medial side can be sacrificed, but all mucous membrane from the lateral flap must be respected. If the operation is morphologically accurate, the result will improve with age. If not morphologically correct, the lip will get worse as the patient grows older (see Plates F and D).

I do not consider it wise to operate on a hare lip under six weeks of age. I prefer between eight and twelve weeks. Foggy weather should be avoided, and the more isolated the baby is from contacts the better. As to feeding whilst awaiting operation, a bottle with a bigger opening than normal in the teat—and flanged—may be used, or spoon feeding. As the latter method is slow and tiring to both baby and nurse, the baby should be fed every two to three hours. Breast milk is best. The babies should be well nourished, given as much sunshine and fresh air as possible, and any anæmia corrected, for anæmia is very common in these patients.

#### OPERATION

The only preliminary medication employed in infants is atropine. Gr. 1/300 is given half an hour before operation. In older children an appropriate dose of secenal  $\frac{3}{4}$  gr. per stone is given along with 1/100 atropine.

*Anæsthesia Employed.*—Intracheal gas, oxygen, and ether. Care should be taken that the intratracheal tube does not become kinked. It is a small tube, and kinking is easy unless a special armoured tube is employed or one specially strengthened. These armoured tubes should be stitched to the back of the tongue as they tend to ride up, due to their spring, and so come out of the trachea during the operation.

#### CLEFT PALATE

Our eyes view a face, but it is only by our ears that a palate is judged. A palate may through operation appear to be anatomically normal, yet, if normal speech is not acquired, the operation is more or less a failure. For normal speech the palate must be capable of shutting off the nasal cavity from the mouth, and it must be soft and muscular, capable of fine vibrating movements, causing inflections of speech. An operation that produces this is a good operation. The operation should cause little distortion of the teeth.

*In Normal Speech.*—When about to speak the superior constrictor becomes tense and the soft palate is raised by the contraction of the levatores palati into a position in contact with the ridge of Passavant. "From this position it is capable of small and rapid movements, the sphincter alternately opening and closing at an extraordinarily high rate."<sup>3</sup>

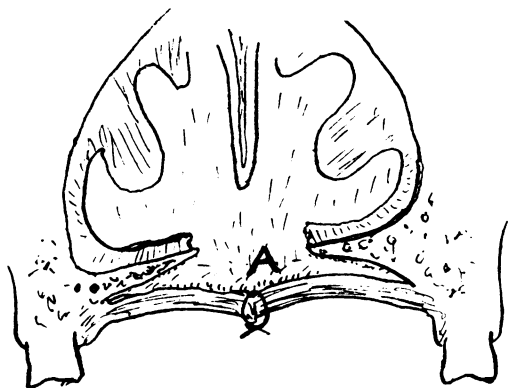
It is seen that for normal speech the soft palate must be capable of closing off the nose and also capable of fine vibratory movements. It is in these two points the classical operations failed.

In 1817 Von Graefe described closure of the soft palate. He caused inflammation of the edges of the cleft and drew them together.

In 1828 Dieffenbach used lateral incisions to ease the tension on the sutural area.

In 1862 Von Langenbeck described his operation. This consisted of freshening the edges of the cleft, dissecting off the mucoperiosteum from the bony palate, and using lateral incisions to allow the flaps to meet in the middle line.

This operation naturally left a raw area on the nasal side and lowered the vault of the palate; the raw area on the nasal side was exposed to nasal bacteria, so that it frequently became infected, and breaking down of suture line occurred frequently, or, at best, a hard fibrosed palate (fig. G).



**Fig. G**

Note raw surface A on nasal side exposing the suture line to infection.

Flaps from the cheek or transposed from a distance, e.g., a finger, have been used to close the cleft, but, as Veau aptly suggests, these operations spring from the same motives as those of Omar, who burnt the Great Library in Alexandria, so that he might be the talk of the learned.<sup>2</sup>

Lane operated before the first dentition and took a flap from across the alveolar ridge, turned the flap like a hinge to lie across the gap. This allowed the baby to suck, but the interference with the alveolar ridge and the subsequent contraction of the scar tissue caused a marked distortion of the alveolar ridge and inequalities of the teeth (Plate H). Brophy, in America, sought to close the cleft by compressing the thin malleable bones of the baby and holding the maxillæ together by a wire suture passed through the maxillæ from one side to the other. This wire, although not interfering with the milk teeth, frequently passed through the permanent teeth buds.

It was following on the results of these operations that dentists drew attention to the severe handicap patients suffered from the irregularities and malocclusion of the teeth (Plate H). Prosthesis in these cases was very difficult, and, after long and tedious sittings, only very unsatisfactory results were obtained. Even speech was unsatisfactory, for the palate was hard and immobile from fibrosis. Fry and

others reasoned that better results would be obtained by dental fitting of plates and velum on patients without any operation on the palate, for the molar teeth would develop normally, and the resulting speech would be equally good. As the child grew, naturally these palate appliances had to be changed. If it had not been for the work of Veau, who developed a new technique and principle of operation, probably we should be still indebted to the dentists for the complete treatment of the cleft palate.

Apart from the distortion of the premolar and molar teeth following on scarring operations, irregularities of the teeth are to be found as an associated defect in all cases of cleft palate where the premaxilla is involved. This defect is usually limited to the lateral incisors, but often the canine is involved. The cleft passes through the region of the lateral incisor, and the latter may lie with the central incisor or be displaced laterally with the canine tooth; a supernumerary lateral incisor may be present. As the premaxilla normally carries the lateral incisor, it seems at first thought strange that in a cleft passing between the premaxilla and the maxilla the lateral incisor should bear such an inconstant position in cleft palate. This must be explained embryologically.

"The union of the globular and maxillary processes occurs between the fifth and sixth week. The primitive dental lamina is formed as a continuous semi-circular ingrowth within the lateral margin of the mandibular arch at the seventh week."<sup>4</sup> The tooth bud is not defined until the seventh week and is invaginated by the mesodermal papillæ from the enamel organ in embryos of the ninth and tenth week. Some think it is the dropping down of the enamel epithelium that prevents the union of premaxilla and maxilla. A missing or poor lateral incisor is supposed to show a hereditary tendency to cleft palate.

It is the distant results that the dentist sees, and, since it is at this stage his services are required, and the surgeon sees the patients only for a short time after the operation and often does not see the late results of his interference. The surgeon frequently has a higher opinion of his work than his colleague the dentist.

One might reckon that the modern operation is a combination of Lane and Langenbeck's operation associated with closure of the mucous membrane on the nasal side and a resuture of the muscles of the soft palate.

For the operation for cleft palate to be wholly successful, i.e., producing a perfectly normal speech, it should be done before the child has got into the habit of producing his speech abnormally. Theoretically, the best time would be in the first year, but the two great objections to this are: (1) the severity of the operation on a young infant, resulting in a high mortality; and (2) the practical difficulty to the surgeon of working in a very small confined space. For these reasons the operation is delayed till the child is between eighteen months and two years old. At this age the mortality is between two per cent. and four per cent. and normal speech is still to be expected after a successful operation without any speech training. The less speech training that is necessary the better, as it is often associated with abnormal grimaces. The early repair of the palate allows the pull of the muscles of both sides to work in conjunction. As the muscles of

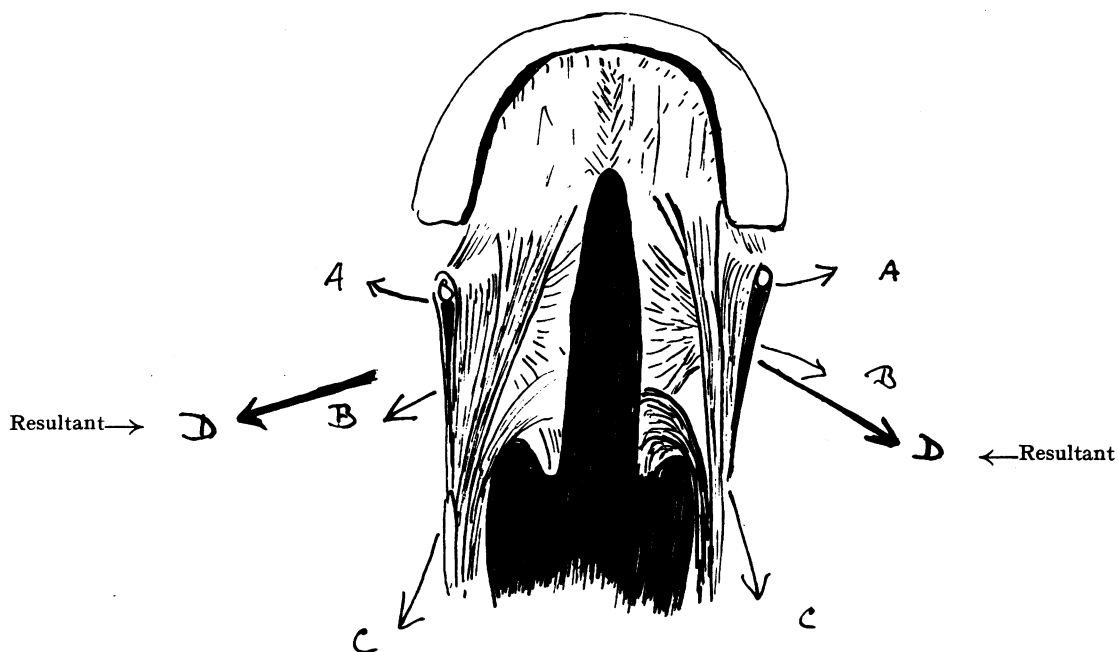
the palate of both sides, acting in conjunction, contract, they pull the palate back and so cause it to lengthen (figs. L1 and L2). This lengthening tends to increase with age. The unrepaired cleft palate is essentially a very short palate (figs. K1 and K2), for hitherto the muscles on each side have been acting separately, and on contracting tend to pull that part of the palate to the side. There is no concerted pull backwards (fig. L1).



**Fig. J**

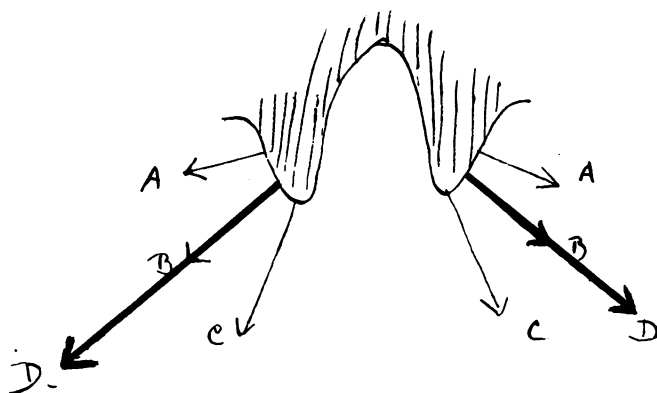
The musculature of the soft palate in a case of cleft palate A compared with the musculature in that of a normal palate B. The muscles of the soft palate in the former are ill developed, and some have taken an abnormal attachment anteriorly to the edge of the palatine bone.

*(After Veau, with kind permission of Messrs. Masson et Cie)*



**Fig. K1**

In a cleft palate the muscles pull in directions A, B, and C, but as C has got some attachment to the post edge of the palate bone it is of little account in pulling back the palate; the resultant force is outwards and not backwards, and the palate tends to remain short.



**Fig. K2**



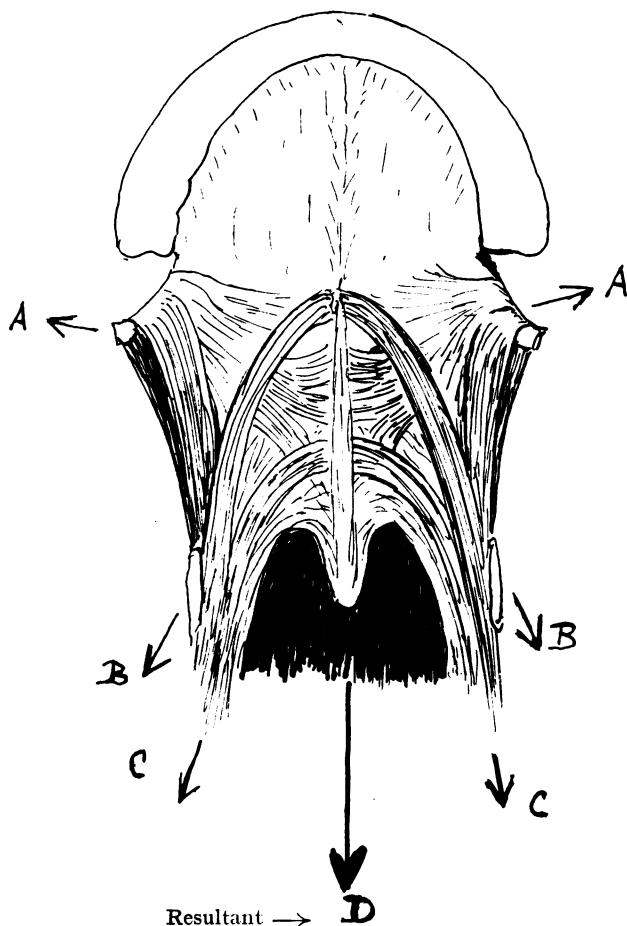


Fig. L1

In the united or normal palate the muscles are better developed and the resultant of the pull of the muscles is straight back (D), so lengthening the palate.

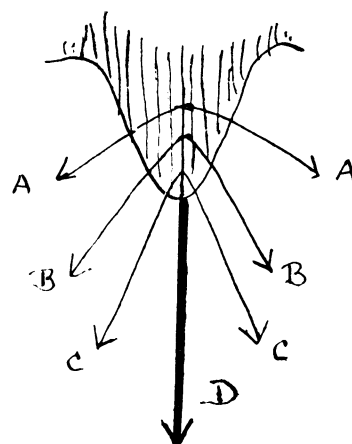


Fig. L2

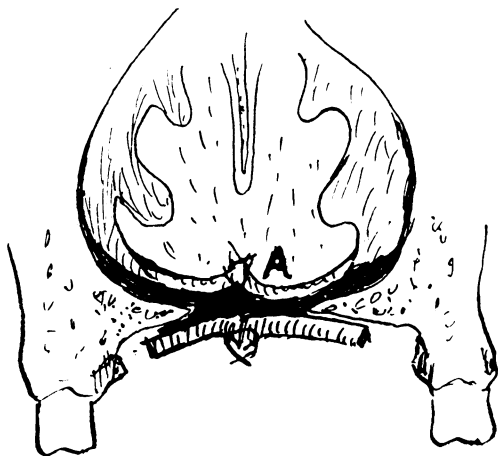
Veau states he was stimulated by Lane's work on cleft palate. In Veau's operation the palate is divided into the nasal mucosal, the muscular, and the palatal mucosal layers. He lays stress on these points :—

1. On the suture of the nasal mucosa of one side to the nasal mucosa of the opposite side of the cleft or in too wide a cleft by using flaps of nasal mucosa from the vomer, in this way leaving no raw surface above to catch nasal drips of infecting secretions.
2. On bringing the muscles of the soft palate (fig. M) together, so causing a muscular palate.
3. On attempting to lengthen the palate by means of sliding flaps. (Wardill followed the same plans, but modified them by making his palatal mucosal

flaps shorter and by attempting to bring the ridge of Passavant forward.)

Denis Browne uses the first two principles of Veau, but does not construct palatal flaps and passes a stitch from the soft palate to behind the constrictor muscles at the ridge of Passavant, which, when tightened, pulls the soft palate back towards this prominence, and at the same time pulls the muscles of the palate of each side together.

From my own experience I am convinced of the necessity of the first two points stressed by Veau, i.e., the closure of the nasal mucosa, and, secondly, the suture of the muscles, but I have found no help in making flaps on the palatal side with a view to lengthening the palate. For, if these flaps are to produce any effect, it would be necessary to have corresponding flaps cut on the nasal side. It is the intact nasal mucosa that prevents these palatal flaps sliding back as theoretically planned and practised by Veau and Wardill. Latterly I have given up cutting these flaps, with no loss to the length of the palate, so that my present practice is to follow more closely the modifications suggested by Denis Browne. Two to three months before the operation on the palate the tonsils are dissected out, any marked adenoid removed, and the post-palatine arteries torn across. The actual operation on the palate consists of separating the muscles of the soft palate from the side walls of the pharynx, breaking the hamular process of the pterygoid, and deepening the separation right to the base of the skull; further forward the mucosa is separated from the rudimentary palate bone on the palate side; the posterior edge of this palate bone is completely freed, and on the nasal side the mucosa is separated from the free medial edge as much as possible. When well mobilised laterally, the two sides of the palate will be touching each other ready for suture (fig. M). There is thus no tension on the sutured palate, and the closure of the nasal surface prevents



**Fig. M**

Note the closure of the nasal mucosa, shutting off droplet infection from the palatal suture line.

infection (fig. M). It is now the greatest rarity for a palate operation to break down. In the older operations break down of the suture line was common.

In this series there were over 100 operations—55 on the palate, and 46 on the lip. There were four deaths :—

One hare lip died following a misadventure the evening following the operation, although it had recovered from the operation.

Three died as the result of the cleft palate operation :—

One died in the early part of the series as the result of a kinked intracheal tube.

One died the evening following the operation. The child was in excellent condition, but started to vomit and suddenly died. This death may have been the result of a small clot in the larynx.

One died on the third day following the operation. The child seemed to be getting on satisfactorily after operation until the third day, when it appeared to be not so well. There was no respiratory distress. It died that evening.

The cause of death was unknown ;

so that there seemed to be two unavoidable deaths. One might estimate the mortality rate for a cleft palate operation about 2 per cent.

The child must be constantly supervised following operation. These children have had to change their method of breathing as the result of the closing of the palate and often find it difficult. In one patient with a short chin the breathing was so difficult that the Denis Browne muscular stitch had to be cut.

There were no complete breakdowns of the palate suture. There were three cases where a small opening showed itself. In one of these closure is expected without further operation. In one, a small anterior perforation was closed at a later date. One has a small hole in the side as the result of experimental cutting across the side flap in a Denis Browne operation, but the anterior blood supply was too poor and a small part of the flap necrosed. This opening does not affect the muscular part of the palate and will easily be closed by a transposed flap.

Speech result is excellent, i.e., normal, in all those operated upon when young. Older children, who had acquired a cleft palate speech before operation, did not gain perfectly normal speech, but their speech showed very great improvement. It is in this class of older children that the collaboration of the speech therapist is most necessary.

If a child has learned to speak before operation he will not just resume normal speech when he wakes up from the anæsthetic, but gradually he should lose his nasal tone, and it may be six to eighteen months before he speaks normally. Occasionally, operation does not produce a competent sphincter, and in these cases special speech training will be needed.

The longer the operation is delayed, the longer will it be before normal speech is attained. The older the child, the more difficult is it to train it to speak normally—ambition and environment are the main factors.

Occasionally a young child may have a normally functioning palatopharyngeal sphincter produced by operation, yet it is unable to use it in speech, and it must be trained.

I am indebted to Messrs. E. and J. Masson et Cie for permission to make copies V.C., E, and J from the illustrations in Mr. Victor Veau's book on "Division Palatine." It is also through the generosity of the editor of *The British Dental Journal* that I can publish Plate H from an article by the late Mr. A. T. Pitts.

I cannot conclude this paper without expressing my gratitude to the anæsthetists, especially Dr. Geddes, for their co-operation in these operations. I should like to say how much the good results and low mortality have been contributed to by the careful nursing and supervision of Sister Mills and the nurses at the Ulster Hospital. Miss Mitchell has been very helpful in training the speech of the older children who had acquired speech defects before their palates had been operated upon.

#### REFERENCES

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2. VEAU, V.: "Division Palatine," Paris: Masson. Cie.; 1938.
3. MORLEY, M.: "Cleft Palate," E. & S. Livingstone; 1945.
4. PITTS, A. T.: "Some Dental Aspects of Cleft Palate," *The British Dental Journal*, May 1, 1922.

#### REVIEWS

**A COMPANION IN SURGICAL STUDIES.** By Ian Aird. Pp. 1068. 1949. Edinburgh: E. & S. Livingstone Ltd. 63s.

THIS should be a very useful book for those taking higher surgical examinations and for surgeons in their clinical teaching. The well-chosen references should make it valuable as a starting-off place for the study of the literature of any particular subject.

"Ian Aird's Notes" were famous in Edinburgh before the war and he published them privately in book form. They made rather difficult reading, as the material they contained was very much condensed and as the headings were few and far between.

The present book is an elaboration of the previous notes, brought up to date very well indeed, and with adequate headings.

Though some of the subjects are dealt with rather briefly, the amount of information given generally is much more than the average text-book contains. S. A. V.

**GAS AND AIR ANALGESIA.** By R. J. Minnitt. Fourth Edition. Pp. vii + 86, with 22 illustrations. 5s. net.

How we welcome a new edition by this indefatigable author. His little book is a delight to handle, as it is excellently printed and illustrated. It is gratifying to realise that there is at least one man in the world who is pegging away to make childbirth easier. One wonders if mothers all over the world ever realise what Dr. Minnitt is doing for them.

He is also doing a great deal for those who lecture to midwives on gas and air analgesia. His little book is an excellent text-book—clear and concise—and is invaluable to those of us who try to explain, in simple language, the workings of the gas and air machines.

We are pleased to see that the author stresses the human element and states that "the attitude of the midwife, or medical attendant, has an important bearing on the amount of relief the patient receives." It is gratifying to note that some emphasis is placed on contradicting the belief that all the anæsthetist has to do is to sit back and twiddle the taps.

This book can be recommended with confidence to all those who are interested in gas and air analgesia. What could be a more fitting ending to this brief review than a quotation from the author? "So may there dawn renewed hope in the hearts of women." V. L.

## Types of Hare Lip

### Plate A



1

Complete facial cleft. Note the alveolus or palate is not affected. (Photograph by kind permission of Mr. C. A. Calvert).



2

Complete unilateral hare lip and cleft palate.



3

Bilateral hare lip and cleft palate. Note how the pre-maxilla is grossly misplaced forward, carrying with it the skin of the probrium, which projects beyond the tip of the nose.



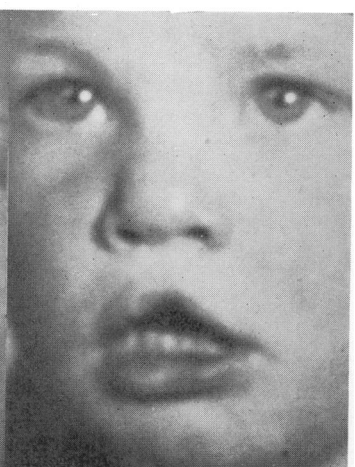
4

Incomplete hare lip. The nostril is not involved.



5

Incomplete hare lip that appears median, but is in reality a left-sided hare lip. Note the unbroken skin with the muscle defect in the region of the defect.

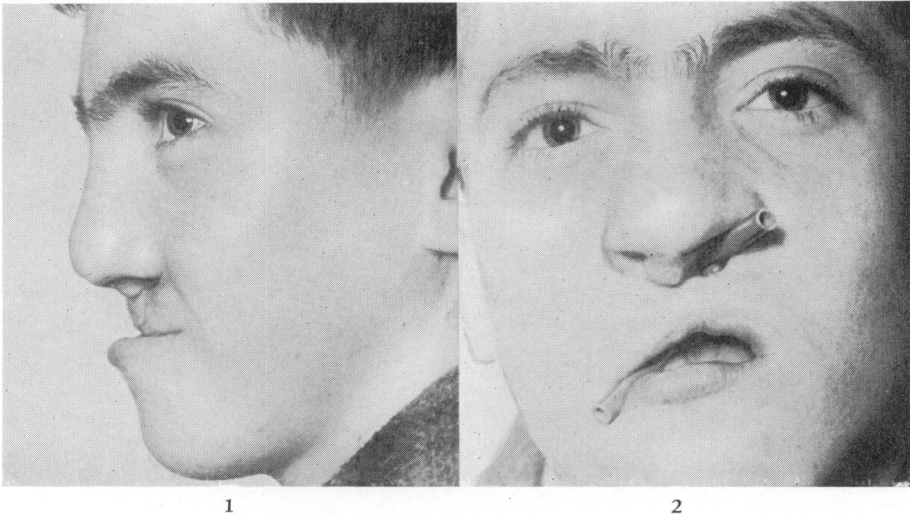


6

A bilateral hare lip. The right side has been repaired, and only then did the left side defect become apparent.

## Late Result of Old Type Operation

**Plate D**



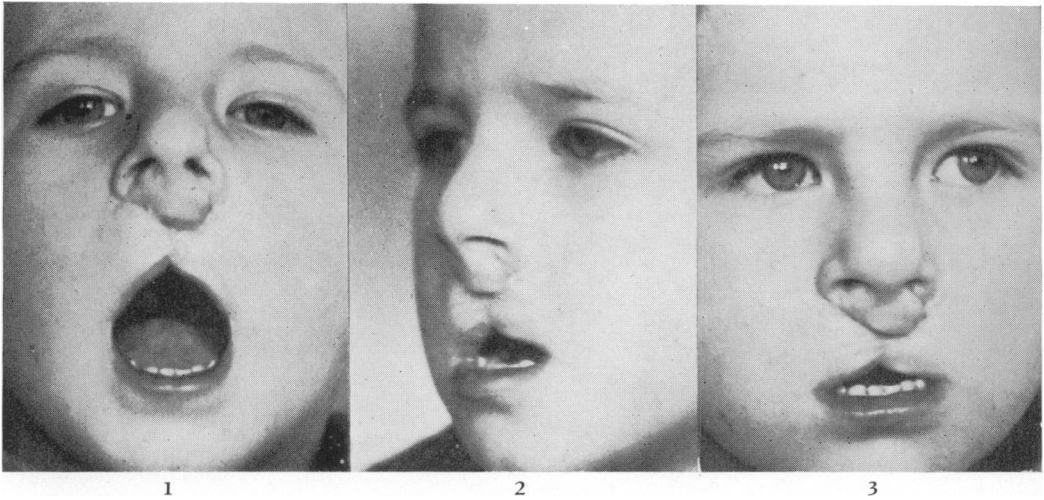
- (1) The operator in the case above has not paid attention to the replacement of the embryonic elements. Note the prolabium displaced under the columella and not brought down to the red margin.

It also shows how, by neglecting to form a floor of the nostril, a sinus exists between the nose and the mouth. No operation performed on the palate will produce good speech till this lip is undone and the floor of the nostril reconstructed.

Note the indrawn upper lip. The upper lip normally extends well beyond the lower lip.

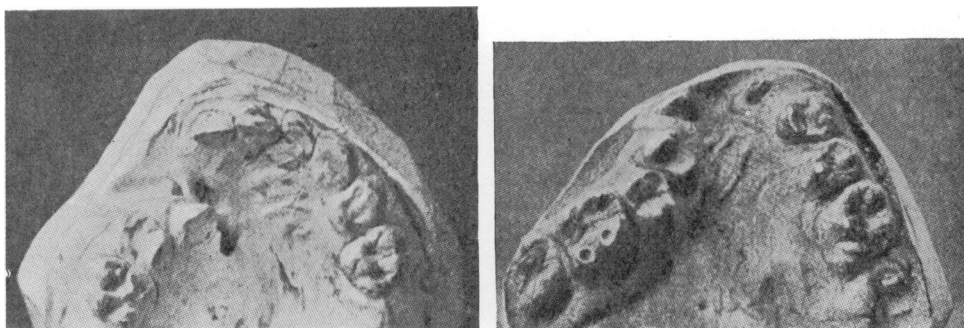
**Late Result of "Non-Embryonic" Plastic Operation**

**Plate F**



This shows the result of a non-embryonic operation. This boy had evidently had a bilateral hare lip and cleft palate. The result immediately after operation may have appeared good, but as growth took place the misplaced embryonic segments have exaggerated the difference from the normal. This child had to have the lip completely undone and rebuilt, bringing the embryonic elements back to their normal position so as natural growth might take place.

**Plate H**



Casts to show deformities of the teeth following on the flap operation of Lane.

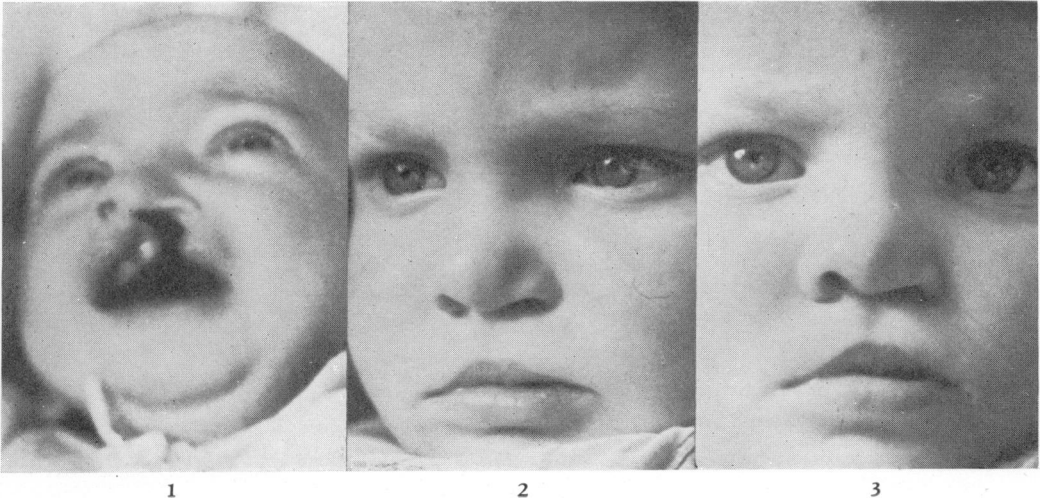
*(By permission of British Dental Journal)*



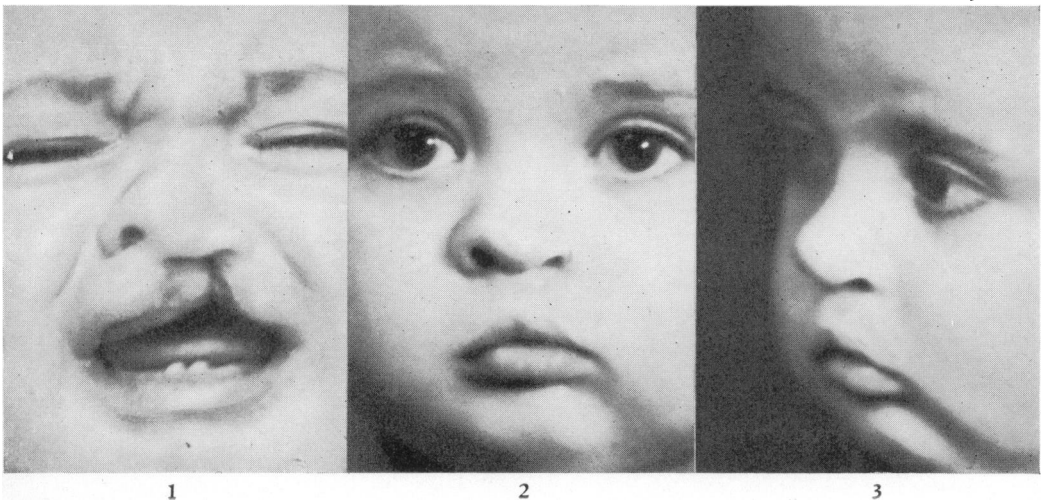
HARE LIP.

THE following illustrations are of complete hare lips with cleft palates which have been now repaired, and speech should be normal. Note the full, thick muscular lips, which project as in the normal beyond the lower lip.

**Plate N**

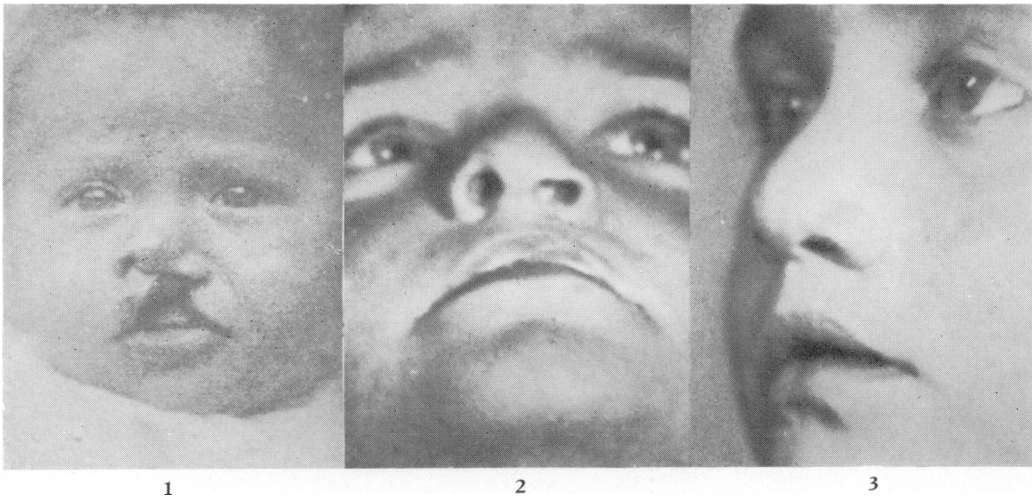


**Plate O**

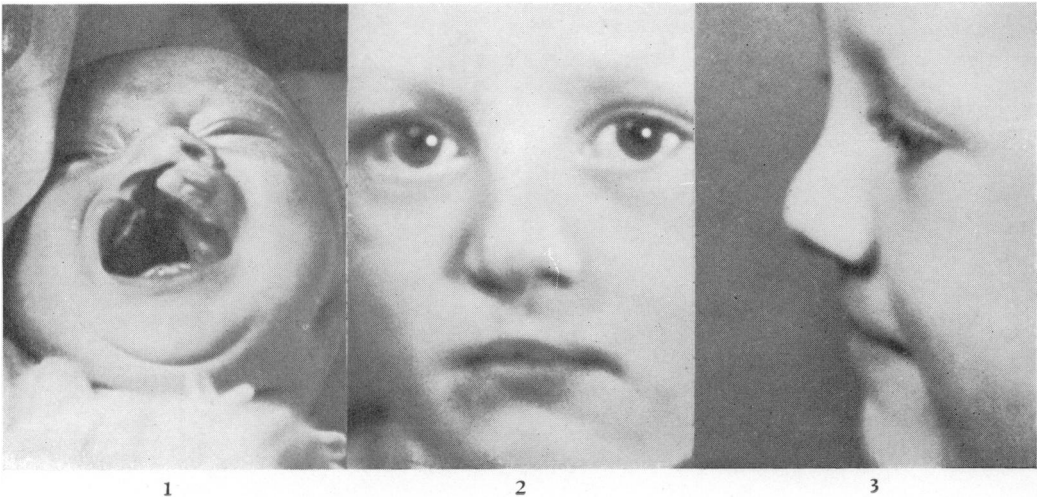


HARE LIP.

**Plate P**



**Plate Q**



**Plate R**



1

2

3

**Plate S**

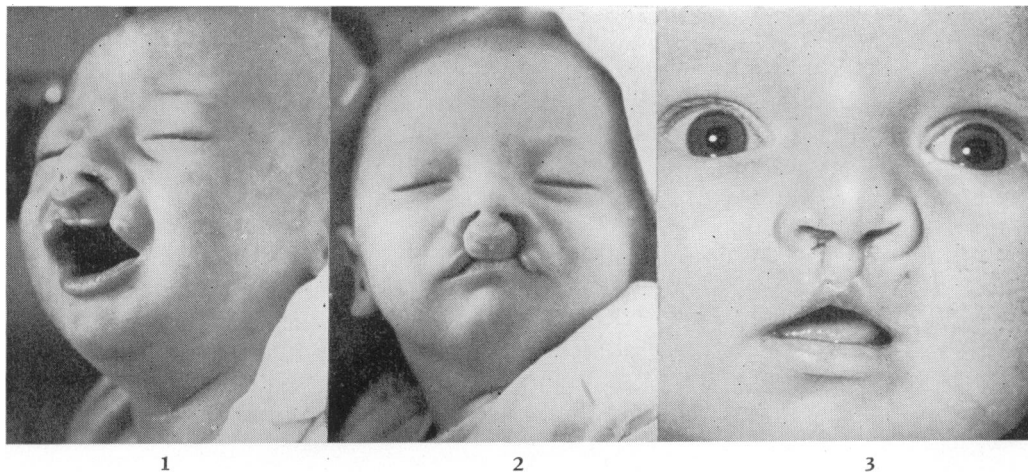


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**Plate T**



(1 and 2) Double hare lip with marked protrusion of the premaxilla.

(3) The same shortly after operation (Denis Browne's). This is too early to determine the late success of this type of operation. This child's appearance two to three years later would be a better indication of the success of the operation.

**Plate U**



These children had had complete unilateral hare lips and cleft palates. Photographs before operation were not taken.